Functional outcomes in children and young people with dyskinetic cerebral palsy

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ABBREVIATIONS

CFCS	Communication Function Classification System
EDACS	Eating and Drinking Ability Classification System
MACS	Manual Ability Classification System
VSS	Viking Speech Scale

AIM This cross-sectional study aimed to map the functional profile of individuals with dyskinetic cerebral palsy (CP), to determine interrelationships between the functional classification systems, and to investigate the relationship of functional abilities with dystonia and choreoathetosis severity.

METHODS Fifty-five children (<15y) and young people (15–22y) (30 males, 25 females; mean age 14y 6mo, standard deviation 4y 1mo) with dyskinetic CP were assessed using the Gross Motor Function Classification System (GMFCS), Manual Ability Classification System (MACS), Communication Function Classification System (CFCS), Eating and Drinking Ability Classification System (EDACS), and Viking Speech Scale (VSS), as well as the Dyskinesia Impairment Scale.

RESULTS Over 50 per cent of the participants exhibited the highest limitation levels in GMFCS, MACS, and VSS. Better functional abilities were seen in EDACS and CFCS. Moderate to excellent interrelationship was found among the classification scales. All scales had significant correlation (r_s=0.65 – 0.81) with dystonia severity except for CFCS in the young people group. Finally, only MACS (r_s=0.40) and EDACS (r_s=0.55) in the young people group demonstrated significant correlation with choreoathetosis severity.

INTERPRETATION The need for inclusion of speech, eating, and drinking in the functional assessment of dyskinetic CP is highlighted. The study further supports the strategy of managing dystonia in particular at a younger age followed by choreoathetosis in a later stage.

Dyskinetic cerebral palsy (CP) accounts for 10 to 15 per cent of CP cases[1] and is characterized by involuntary, uncontrolled, and stereotyped movements.[2] In dyskinetic CP the dominant movement disorders dystonia and choreoathetosis[2,3] are often simultaneously present.[4] Dystonia is described as involuntary sustained or intermittent muscle contraction causing twisting and repetitive movements, abnormal postures, or both. Chorea refers to rapid, involuntary, jerky, often fragmented movements; while athetosis means slower, constantly changing, writhing, or contorting movements.[5,6]

For appropriate management of individuals with CP, functional classification systems play an important role because they allow a methodical approach in distinguishing characteristics of functional abilities.[7] Functional classification systems also aid in prognostication, goal setting, and management planning, according to the study of Rosenbaum et al.[7] Currently the most commonly used CP classification scales are the Gross Motor Function Classification System (GMFCS),[8] the Manual Ability Classification System (MACS),[9] and the Communication Function Classification System (CFCS).[10] The Eating and Drinking Ability Classification System (EDACS)[11] and the Viking Speech Scale (VSS)[12] are two more recent scales. Further, despite dyskinetic CP being the second largest CP group,[13] few studies have focused on functional classification in this population. However, a comprehensive functional profile of dyskinetic CP and its relationship with dystonia and choreoathetosis severity is vital in establishing targeted interventions.

In 2012, Hidecker et al.[14] first reported a moderate to good correlation between the gross motor, manual, and communication abilities of children and young people with CP. However, the lack of information on the percentage of the sample having dyskinetic CP requires caution in generalizing the findings to the population of dyskinetic CP. Identifying the subtype of CP in terms of predominant motor characteristics is valuable as nature of movement disorder may impact on functionality in a distinct way.[15] More recently, Elze et al.[16] investigated the association

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between dystonia and GMFCS, MACS, and CFCS in children with childhood dystonia disorders, where dyskinetic CP represented 47 per cent of the sample. The results showed a strong relationship between dystonia and the three classification systems. The association between choreoathetosis and the three functional scales was not investigated. Strong interrelationship was found among the GMFCS, MACS, and CFCS. Despite the larger representation of dyskinetic CP in this study, the GMFCS and MACS levels I and II were underrepresented, whereas level V was overrepresented, in comparison with previous dyskinetic CP population studies.[1,17] Therefore a more focused approach is warranted for this target population. Another recent study[4] used the Dyskinesia Impairment Scale (DIS)[18] in individuals with dyskinetic CP, whereby both dystonia and choreoathetosis could be specifically investigated. High correlation was found between dystonia and the GMFCS, MACS, and CFCS. Conversely, no significant relationship was found with choreoathetosis.

In these studies,[4,16] gross motor, manual ability, and communication skills were investigated but other vital daily functions such as speech, eating, and drinking were not included by means of the more recent classification of EDACS and VSS. Establishing the relationship of dystonia and choreoathetosis with speech and eating and drinking abilities is essential to broaden the functional profile of individuals with dyskinetic CP.

This study therefore specifically aims: (1) to map the functional profile of individuals with dyskinetic CP by using five functional classification scales including the recent EDACS and VSS; (2) to determine the relationship between the functional classification scales; and (3) to investigate the relationship of the different functional classification systems with dystonia and choreoathetosis. It is hypothesized that individuals with dyskinetic CP are represented in the most severe functional classification levels.

METHODS

Participants

Children and young people from five Flemish special education schools for motor disabilities were screened for inclusion in this study. Individuals aged 5 to 22 years with dyskinetic CP and able to understand test instructions were eligible to participate. Based on the universal definition established by the United Nations,[19] participants were classified either in the children (<15y) or young people group (15–22y). Individuals who underwent orthopaedic or neurosurgical interventions within the previous year and spinal fusion were excluded. The study was approved by the Ethical Committee of KU Leuven. Written informed consent was obtained from the parents and participants.

Assessment

Gross motor function, manual ability, eating and drinking functions, communication, and speech were assessed using the corresponding classification systems of GMFCS, MACS, EDACS, CFCS, and VSS. All systems utilize a five-level ordinal rating scale with higher levels indicating more severe functional disability, except the VSS with a four-level ordinal rating. The classification systems have been proven to be reliable in individuals with CP.[8–12] The definitions of the respective functional levels are presented in Table SI (online supporting information).

To assess dystonia and choreoathetosis severity, the DIS[17] was administered. The DIS is a reliable and valid video-based ordinal scale and is subdivided into dystonia and choreoathetosis subscales. They distinctly assess dystonia and choreoathetosis of the eyes, mouth, neck, trunk, and extremities at rest and during activity.[4,18,20] Total dystonia and choreoathetosis scores are expressed as percentages.

What this paper adds

- Dystonia is significantly related with all classifications, except the Communication Function Classification System (CFCS).
- Eating and Drinking Ability Classification System (EDACS) and Viking Speech Scale are significantly related with Gross Motor Function Classification System (GMFCS), Manual Ability Classification System (MACS), and CFCS.
- No significant relationship between choreoathetosis and classification scales at young age.
- Higher relationship between choreoathetosis and MACS and EDACS at older age.

Procedure

One assessor (EM) videotaped all participants with the DIS video protocol in their natural environment at the special education school. Evaluation of functional levels was done on the same day as the video recording while the scoring of dystonia and choreoathetosis levels was performed in the succeeding months.

Statistical analysis

Shapiro–Wilk tests indicated that most variables were not normally distributed. Consequently, non-parametric statistics were used. Descriptive statistics were calculated. Spearman's rank correlation coefficients (r_s) were used to assess the interrelationship between the functional classification systems, and between the classification scales and dystonia and choreoathetosis percentages. In addition, the Wilcoxon signed-rank test was applied to compare distribution of the scales. Box plots were used to visualize dystonia and choreoathetosis severity within the different functional levels. Because of the large age range, data were stratified by two age groups (<15y, 15–22y) and correlation coefficients were compared by using Fisher's z-transformation.

Correlation coefficients above 0.75 were considered as excellent; between 0.50 and 0.75 as moderate to good; between 0.25 and 0.50 as fair; and between 0 and 0.25 as no relationship.[21] Statistical significance was set at $p<0.05$. Data were analyzed using the Statistica version 12 (StatSoft Inc., Tulsa, OK, USA) and Vassarstats[22] for comparison of correlations.

RESULTS

Functional profile

Fifty-five participants (30 males, 25 females) between 5 years and 22 years (mean age 14y 6mo, standard deviation

4y 1mo) fulfilled the criteria. Individual characteristics are provided in Tables SII and SIII (online supporting information). Twenty-five participants were younger than 15 years, while thirty were aged between 15 years and 22 years. Participants were sampled across all levels of the classification systems (see Fig. 1). Ten participants were classified as GMFCS level I; 5 as level II; 6 as level III; 7 as level IV; and 27 as level V. For MACS, four participants were evaluated as level I; eight as level II; seven as level III; 12 as level IV; and 24 as level V. Meanwhile, six participants were categorized as CFCS level I; 20 as level II; 18 as level III; eight as level IV; and three as level V. For EDACS, nine participants were classified as level I; 18 as level II; 10 as level III; 12 as level IV; and six as level V. Eight participants were evaluated as VSS level I; 12 as level II; 11 as level III; and 24 as level IV.

Relationship between functional classification scales

The association between the functional classification scales are presented in Table SIV (online supporting information). Excellent relationship (p<0.001) was found between GMFCS and MACS (r_s=0.91; 95% confidence interval [CI] 0.85–0.95), EDACS (r_s=0.78; 95% CI 0.65–0.87), and VSS (r_s=0.79; 95% CI 0.66–0.87), and between MACS and EDACS (r_s=0.77; 95% CI 0.64–0.86). Further, a moderate to good relationship (p<0.001) was found between GMFCS and CFCS (r_s=0.54; 95% CI 0.32–0.70), MACS and VSS (r_s=0.73; 95% CI 0.58–0.83), CFCS and VSS (r_s=0.51; 95% CI 0.28–0.68), and EDACS and VSS (r_s=0.73; 95% CI 0.58–0.83). Finally, a fair relationship (p<0.001) was demonstrated for CFCS and EDACS (r_s=0.49; 95% CI 0.26–0.67), and MACS and CFCS (r_s=0.46; 95% CI 0.22–0.65).

Cross-tabulations of the GMFCS, MACS, EDACS, and CFCS are presented in Table I. Matching levels of functional classification were observed mostly between GMFCS and MACS, especially for those with higher functional severity. Matching levels were least observed between MACS and CFCS. Wilcoxon signed-rank tests showed significant differences (p<0.001) in the distribution between the scales, except for GMFCS and MACS (p=0.170), and for EDACS and CFCS (p=0.462).

Relationship between dystonia and choreoathetosis and functional classification scales

The box plots of dystonia and choreoathetosis percentages for the respective levels of GMFCS, MACS, EDACS, CFCS, and VSS are presented in Figure S1 (online supporting information). The box plots illustrate the relationship of dystonia and choreoathetosis with the functional level of the participants.

In the total sample, all functional classification scales were found to have a significant relationship (p<0.001) with dystonia. Conversely, only EDACS was significantly associated (p=0.039) with choreoathetosis. For dystonia, a moderate to good relationship (r_s=0.61–0.70; 95% CI 0.41–0.82) was found with all functional classification scales, except for a fair association with the CFCS (r_s=0.36; 95% CI 0.11–0.57). For choreoathetosis, a fair relationship was found with the EDACS (r_s=0.28; 95% CI 0.16–0.51) and CFCS (r_s=0.26; 95% CI –0.50 to 0.01) but not with the other functional classification scales.

Because of the wide age range, the results of the two age groups were differentiated into a children group and a young people group in accordance with UN definitions.[19] Table SV (online supporting information) summarizes the relationship of the functional classification scales with dystonia and choreoathetosis scores stratified by age.

Children (<15y)

All functional classification scales were significantly associated with dystonia percentage scores. An excellent

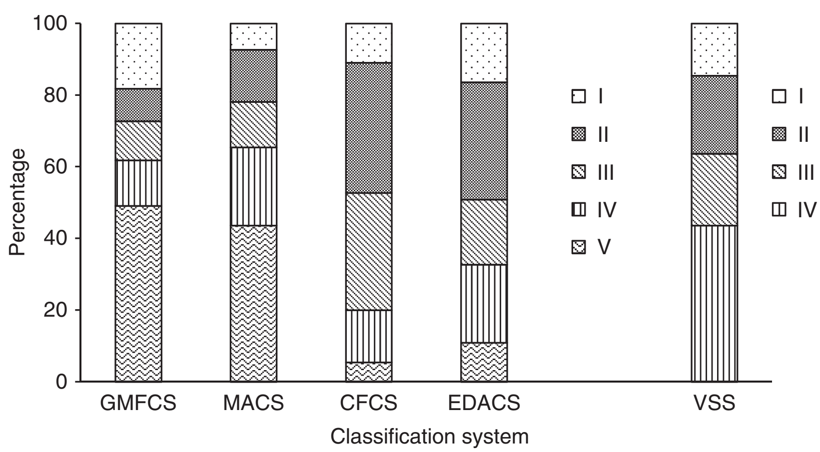

Figure 1: Distribution of functional levels by classification system in 55 individuals with dyskinetic cerebral palsy. GMFCS, Gross Motor Function Classification System; MACS, Manual Ability Classification System; CFCS, Communication Function Classification System; EDACS, Eating and Drinking Ability Classification System; VSS, Viking Speech Scale

Table I: Cross-tabulation of GMFCS, MACS, EDACS, and CFCS for 55 participants (%)

	GMFCS level					
MACS level	I	II	III	IV	V	Total
I	4 (7)	0 (0)	0 (0)	0 (0)	0 (0)	4 (7)
II	5 (9)	2 (4)	1 (2)	0 (0)	0 (0)	8 (15)
III	0 (0)	3 (5)	3 (5)	1 (2)	0 (0)	7 (13)
IV	1 (2)	0 (0)	2 (4)	5 (9)	4 (7)	12 (22)
V	0 (0)	0 (0)	0 (0)	1 (2)	23 (42)	24 (44)
Total	10 (18)	5 (9)	6 (11)	7 (13)	27 (49)	55 (100)

	GMFCS level					
EDACS level	I	II	III	IV	V	Total
I	8 (15)	0 (0)	1 (2)	0 (0)	0 (0)	9 (16)
II	2 (4)	4 (7)	3 (5)	5 (9)	4 (7)	18 (33)
III	0 (0)	1 (2)	1 (2)	2 (4)	6 (11)	10 (18)
IV	0 (0)	0 (0)	1 (2)	0 (0)	11 (20)	12 (22)
V	0 (0)	0 (0)	0 (0)	0 (0)	6 (11)	6 (11)
Total	10 (18)	5 (9)	6 (11)	7 (13)	27 (49)	55 (100)

	GMFCS level					
CFCS level	I	II	III	IV	V	Total
I	6 (11)	0 (0)	0 (0)	0 (0)	0 (0)	6 (11)
II	1 (2)	3 (5)	4 (7)	5 (9)	7 (13)	20 (36)
III	3 (5)	2 (4)	1 (2)	2 (4)	10 (18)	18 (33)
IV	0 (0)	0 (0)	1 (2)	0 (0)	7 (13)	8 (15)
V	0 (0)	0 (0)	0 (0)	0 (0)	3 (5)	3 (5)
Total	10 (18)	5 (9)	6 (11)	7 (13)	27 (49)	55 (100)

	MACS level					
EDACS level	I	II	III	IV	V	Total
I	4 (7)	5 (9)	0 (0)	0 (0)	0 (0)	9 (16)
II	0 (0)	3 (5)	5 (9)	6 (11)	4 (7)	18 (33)
III	0 (0)	0 (0)	1 (2)	5 (9)	4 (7)	10 (18)
IV	0 (0)	0 (0)	1 (2)	1 (2)	10 (18)	12 (22)
V	0 (0)	0 (0)	0 (0)	0 (0)	6 (11)	6 (11)
Total	4 (7)	8 (15)	7 (13)	12 (22)	24 (44)	55 (100)

	MACS level					
CFCS level	I	II	III	IV	V	Total
I	3 (5)	2 (4)	0 (0)	1 (2)	0 (0)	6 (11)
II	1 (2)	1 (2)	5 (9)	6 (11)	7 (13)	20 (36)
III	0 (0)	5 (9)	1 (2)	4 (7)	8 (15)	18 (33)
IV	0 (0)	0 (0)	1 (2)	0 (0)	7 (13)	8 (15)
V	0 (0)	0 (0)	0 (0)	1 (2)	2 (4)	3 (5)
Total	4 (7)	8 (15)	7 (13)	12 (22)	24 (44)	55 (100)

	EDACS level					
CFCS level	I	II	III	IV	V	Total
I	5 (9)	1 (2)	0 (0)	0 (0)	0 (0)	6 (11)
II	1 (2)	9 (16)	5 (9)	4 (7)	1 (2)	20 (36)
III	3 (5)	7 (13)	4 (7)	2 (4)	2 (4)	18 (33)
IV	0 (0)	0 (0)	1 (2)	5 (9)	2 (4)	8 (15)
V	0 (0)	1 (2)	0 (0)	1 (2)	1 (2)	3 (5)
Total	9 (16)	18 (33)	10 (18)	12 (22)	6 (11)	55 (100)

GMFCS, Gross Motor Function Classification System; MACS, Manual Ability Classification System; EDACS, Eating and Drinking Ability Classification System; CFCS, Communication Function Classification System.

relationship was observed for the GMFCS (r_s=0.79), a moderate to good relationship for the MACS (r_s=0.72), EDACS (r_s=0.52), and VSS (r_s=0.60), and a fair relationship for the CFCS (r_s=0.49). Conversely, no significant association was found between the functional classification scales and choreoathetosis.

Young people (15–22y)

All functional classification scales, except CFCS, were significantly correlated with dystonia percentage scores. Specifically, excellent relationship was found between dystonia and the EDACS (r_s=0.81) while a moderate to good relationship was demonstrated for GMFCS (r_s=0.68), MACS (r_s=0.71), and VSS (r_s=0.65).

For choreoathetosis, no significant association was found with GMFCS, CFCS, and VSS. However, a moderately significant relationship was found between choreoathetosis percentages and EDACS (r_s=0.55) and a fair relationship (r_s=0.40) for the MACS.

Statistical testing of the differences between the correlation coefficients in both age groups (see Table II) revealed no significant difference, except between EDACS and choreoathetosis (p=0.018).

DISCUSSION

This study was aimed at advancing our insights in the functional profile of individuals with dyskinetic CP and to determine the relationship of dystonia and choreoathetosis with the functional classification systems assessing gross motor ability, upper extremity function, eating and drinking ability, communication, and speech.

The first aim was to map the functional profile of individuals with dyskinetic CP by using five functional classification scales. Cross-tabulations showed a homogeneous distribution, except for GMFCS and MACS, where 42 per cent of the participants were classified as level V. More than 50 per cent of the participants exhibited highest limitation levels not only in gross motor function and manual ability but also in speech production. Conversely, participants showed better functional abilities in communication and eating and drinking functions (levels I–III). In general, the GMFCS and MACS findings correspond with previous results of Himmelmann et al.[1,17] where 79 per cent of the participants were categorized as GMFCS levels IV and V. Severe manual impairment was noted in 77 per cent of the participants. The predominance of GMFCS and MACS levels IV and V also corresponds with the findings of Hidecker et al.[14] Recently Elze et al.[16] also reported similar findings in a group of children with childhood dystonia (including dyskinetic CP), where 68 per cent were classified as GMFCS levels IV and V, and 70 per cent as MACS levels IV and V. For communication, the latter study reported 45 per cent of the participants as CFCS levels IV and V. This proportion is higher than the finding of our current study, noting only 20 per cent of participants categorized in the most severe functional classes of CFCS. The difference could be attributed to the patient population of Elze et al.[16] including different pediatric childhood dystonia conditions. Regarding eating, drinking, and speech production, no previous study assessed the EDACS and VSS in dyskinetic CP. The better abilities in eating and drinking functions compared with gross motor and upper extremity functions is a unique finding and in line with the previously described communication patterns in dyskinetic CP.[4] However, speech production does not follow the EDACS and CFCS patterns. This may be explained by the assessed component of the scales. Communication function and eating and drinking abilities are underpinned by motor processes. However, the focus of the EDACS and CFCS is to describe an individual's ability to safely eat and drink, and to engage in daily communication function regardless of the communication method used. Speech production measured by the VSS focuses on breathing control, phonation, and articulation. From this perspective, the five classification scales are very complementary to profile the upper and lower motor function, as well as oromotor function along with communication and eating and drinking abilities.

Table II: Significance of difference between correlation coefficients (r_s) of the functional classification scales with dystonia and choreoathetosis scores stratified by age

	Dystonia		Choreoathetosis	
	z	p	z	p
GMFCS	0.84	0.401	–0.61	0.271
MACS	0.07	0.944	–1.54	0.124
EDACS	–1.92	0.055	–2.36	0.018
CFCS	0.98	0.327	–0.98	0.164
VSS	–0.29	0.772	–0.88	0.379

z, Fisher's r-to-z transformation; GMFCS, Gross Motor Function Classification System; MACS, Manual Ability Classification System; EDACS, Eating and Drinking Ability Classification System; CFCS, Communication Function Classification System; VSS, Viking Speech Scale.

The second aim was to investigate the interrelationship between the functional classification scales. Moderate to good relationship was found among the GMFCS, MACS, and CFCS which corresponds to the results of Hidecker et al.[14] and Elze et al.[16] The generally higher associations of the current study may be attributed to the differences in the composition of the sample between the studies. For example, the sample of Hidecker et al.[14] comprised participants across the population of CP. No distinction between motor characteristics was applied. Another study of Beckung and Hagberg,[23] involving participants with CP of which 11 per cent had dyskinetic CP, demonstrated that gross motor ability was strongly related to bimanual function. In line with previous studies and taking into consideration that we found no significant difference (p=0.167) between the GMFCS and MACS and the excellent correlation (r_s=0.91) between the two classifications, it is inferred that the GMFCS and MACS are predictive for one another. Further, the current study is the first to additionally determine the relationship of the EDACS and VSS with the GMFCS, MACS, and CFCS in dyskinetic CP. The significant difference in distribution and the fair to moderate relationship between CFCS, VSS, and EDACS with the two scales assessing gross motor ability and manual function underline the need to include communication, speech, eating, and drinking and as a part of the functional assessment in individuals with dyskinetic CP. The non-significant difference in the distribution between EDACS and CFCS also underscores the inverse relationship of communication with motor tasks such as eating and drinking.[24] Current findings further support that individuals with high functional severity in performing motor activities may demonstrate good communication ability.[25] Communication encompasses motor and social components enabling performance of roles as sender and receiver of messages to facilitate interaction. Knowledge of functional profiles can facilitate goal setting and treatment planning by the clinical team, and optimize outcomes of targeted assessment and intervention.

The final aim was to determine the relation of both dystonia and choreoathetosis with the different functional classification systems. Differentiation of dystonia and choreoathetosis severity from functional activities facilitates the analysis of the interrelationship of body function impairments and activity limitations as expressed by the different classification systems. A recent study demonstrated that dystonia had a higher impact on activity, participation, and quality of life than choreoathetosis had.[26] Previous research showed evidence for independent occurrence of dystonia and choreoathetosis in dyskinetic CP[4] and a good relationship was found between dystonia severity and GMFCS, MACS, and CFCS[4,16] but not for choreoathetosis.[4]

In the present study similar findings were found for the EDACS and VSS. Correlation coefficients showed that dystonia increased significantly with increasing levels of the EDACS and VSS. Also for choreoathetosis, no significant correlation was found with the VSS, and only fair relationship with the EDACS. However, in previous studies, a wide age range was used. Therefore, in our study, differentiation has been made between a children group (<15y) and a young people group (15–22y). Results showed no significant difference in the correlation coefficients between both age groups except for the EDACS and choreoathetosis score. Choreoathetosis was significantly associated to EDACS only in the young people group. Although there was only a significant difference in the correlation for EDACS, an interesting result was the significant relationship of MACS and choreoathetosis in the young people group. The increase in association between choreoathetosis and MACS and EDACS in the young people group may be attributed to daily life activities that are performed at this age such as handling and self-care. This is supported by the findings of Rosenbaum et al.,[7] demonstrating that the MACS had a predictive role in the development of self-care. Thus, independence in manual functions is observed in later age. Both findings stress the more prominent role of choreoathetosis at older age for specific functional areas.

In contrast, a different pattern was found for the relation between communication and dystonia. CFCS and dystonia were significantly related in the children group but not in the young people group. This may imply that the impact of dystonia on communication is more prominent during childhood. It is hypothesized that other forms of communication, such as augmentative and alternative communication, are adapted as in individual ages, thus minimizing the impact of dystonia. The impact of personal and environmental factors on the communication ability of individuals with dyskinetic CP also plays a role but needs further investigation. No other studies have addressed the possible influence of age on functional abilities and its relation to the underlying dyskinetic symptoms. To confirm our findings, longitudinal studies that involve larger samples are warranted to determine the natural history of dystonia and choreoathetosis, and their impact on the adaptations involved in the performance of various functions in individuals with dyskinetic CP.

This is the first study, to our knowledge, presenting the full functional profile of individuals with dyskinetic CP, also including the areas of speech, eating, and drinking by using validated classification systems, in addition to gross motor, manual ability, and communication classification. Moreover, the association with both dystonia and choreoathetosis was assessed over a large age span. However, this study also has some limitations. First, although a representative and large cohort of individuals with dyskinetic CP from Flemish schools was evaluated, a larger sample would be needed to further explore age differences in relation to dystonia and choreoathetosis severity. Considering the lower prevalence of dyskinetic CP, an international collaborative study would be recommended to increase the number of participants. This endeavour may increase insight in the influence of other characteristics such as personal factors, and cultural and environmental context on the relationship of the functional classification systems with dystonia and choreoathetosis. Secondly, the effects of age were explored in a cross-sectional design and therefore results should be interpreted with caution. Longitudinal research is needed to confirm our findings. Deepening our insights in developmental changes is crucial for the prognosis of the condition and better management of these patients. Finally, comparison between the associations of the current study with that of other subtypes of CP is not plausible. It would be good to further analyse through a larger study the relationship between the functional classification systems among the subtypes of CP based on predominant motor characteristics.

In conclusion, the study demonstrated that individuals with dyskinetic CP exhibited mostly very severe limitations in gross motor function, manual ability, and speech production, and better abilities in eating, drinking, and communication. The GMFCS, MACS, EDACS, CFCS, and VSS were also found to have a significant interrelation, though of varying levels. Further, dystonia showed significant correlation with the different functional scales across age groups in general. Still, our results suggest that choreoathetosis may play a more important role in for manual ability and eating and drinking functions at older age.

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SUPPORTING INFORMATION

The following additional material may be found online:

Figure S1: Median and interquartile range of the dystonia and choreoathetosis levels for the levels of the Gross Motor Function Classification System (GMFCS), the Manual Ability Classification System (MACS), the Eating and Drinking Ability Classification System (EDACS), the Communication Function Classification System (CFCS), and the Viking Speech Scale (VSS).

Table SI: Definitions of the functional levels of the different classification systems

Table SII: Participant characteristics

Table SIII: Median, interquartile range, and range of the outcome measures on the Dyskinesia Impairment Scale

Table SIV: Intercorrelation coefficients of the functional classification systems

Table SV: Correlation coefficients (r_s) of the functional classification scales with dystonia and choreoathetosis scores stratified by age

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